Neuro-ophthalmology review

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Neuro-ophthalmology deals with visual problems caused by disorders of the brain or the optic nerve connection.
Part 1:
Pupillary Disorders
Anatomy and physiology:

- The pupil size is controlled by a balance between parasympathetic innervation to the sphincter muscles and sympathetic innervation of the dilator muscles of the iris.

- Pupil constricts to light and near stimuli.
Pupillary Disorders

- **Sympathetic (adrenergic) pathway:**
  - Pupillary dilation is mediated through sympathetic (adrenergic) pathway that originate in the hypothalamus
Pupillary Disorders

- parasym pathetic (cholinergic) pathway:

1. Light is shined on right eye only.

2. Action potentials from right eye reach both right and left pretectal nuclei.

3. The pretectal nuclei stimulate both sides of the Eddinger–Westphal nucleus even though the light was perceived only in the right eye.

4. The right and left sides of the Eddinger–Westphal nuclei generate action potentials through the right and left oculomotor nerves, causing both pupils to constrict.

Pupillary Disorders

- Examination of the pupil:
  - Best conducted in dim light room using a bright light
  - The patient should be relaxed and fixing on a distant object
  - The size, shape, and position of each pupil should be noted in light and dark conditions
  - Check light reflex looking for a relative afferent pupillary defect (RAPD)
Pupillary light reflex

http://casemed.case.edu/clerkships/neurology/NeurLrngObjectives/light%20reflex.jpg
Pupillary Disorders

Anisocoria
Which pupil is abnormal?
When the small pupil does not dilate as well as the large pupil in dim light, then the small pupil is abnormal.

When the larger pupil does not constrict as well as the small pupil in response to a light stimulus, then the large pupil is abnormal.
The large pupil is abnormal

- Previous ocular surgery
- Ocular trauma
- Use of medication like cycloplegics e.g. atropine, cyclopentolate
- Third nerve palsy
- Tonic pupil (Adie's pupil)
Sluggish, segmental pupillary responses to light
better response to near followed by slow redilation.
Young female
Unilateral (80%)
Instillation of weak cholinergic agents (0.1% pilocarpine) will cause constriction of the tonic pupil (denervation hypersensitivity)
Benign condition
Tonic pupil (Adie's pupil)

Holmes-Adie syndrome:
- includes tonic pupil, diminished deep tendon reflexes and orthostatic hypotension.
The small pupil is abnormal:

- Previous ocular surgery
- Ocular trauma or inflammation
- Use of medication e.g. pilocarpine
- Horner syndrome
The small pupil is abnormal:

- **Horner syndrome:**
  - Small pupil (miosis), ptosis and anhydrosis
  - Caused by a lesion anywhere along the sympathetic pathway
  - Carotid dissection, carotid aneurysm and tumor can be associated with this syndrome
Part 2:

Neuromotility disorders
### Anatomy and physiology:

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Neuromotility disorders

- Anatomy and physiology:

• 65 yrs old presented to ER complaining of double vision
Third cranial nerve (oculomotor)palsy:
Third cranial nerve (oculomotor)

• Begins as a nucleus in the midbrain that consists of several subnuclei.
• Innervate the individual extraocular muscles, the eyelids, and the pupils
Third cranial nerve (oculomotor)palsy:

**Check for pupil involvement**

Absence of pupillary involvement suggests a benign process that can be observed over a couple of weeks. A fixed, dilated pupil requires extensive neurologic evaluation.
Third cranial nerve (oculomotor)palsy:

*Check for pupil involvement*
Third cranial nerve (oculomotor)palsy:

- **Etiology:**
  - Micro-vascular ischemia (DM and HTN)
  - Intracranial aneurysm (posterior communicating artery)
  - Trauma
  - Brain tumor
Neuromotility disorders

- Fourth cranial nerve (trochlear) palsy:
  - Vertical diplopia
  - Head tilt to the opposite shoulder
Fourth cranial nerve (trochlear) palsy:

- Etiology:
  - trauma
  - idiopathic
  - congenital
Neuromotility disorders

- Which muscle is affected?
Sixth cranial nerve (abducens)palsy:

- Horizontal diplopia (worse at distance)
- Esotropia
- Face turn in the direction of the paralyzed muscle
- Limited Abduction on the side of the lesion
Neuromotility disorders

- Sixth cranial nerve (abducens)palsy:
  - causes:
    - intracranial tumors
    - trauma
    - microvascular diseases
    - increased intracranial pressure
Part 3: Neuromuscular disorder
Ocular myasthenia gravis

- Chronic autoimmune disease affecting the neuromuscular junction in skeletal muscles.
- Ptosis
- Diplopia
- Fatigability and variability of clinical findings are characteristic
- The pupil is not affected
- Check for systemic weakness, difficulty in swallowing or breathing.
- Assess orbicularis strength
- Blood test for acetylcholine receptor antibodies
Ocular myasthenia gravis:

- Tensilon test: inhibits acetylcholinesterase and can transiently reverse signs of weakness due to OMG, such as ptosis and extraocular muscle paresis.
Part 4: Visual pathway disorders
Lesions anywhere in the visual pathway will produce visual field defect
Optic nerve diseases:

- Usually unilateral
- Afferent pupillary defect
- Central visual loss
- Loss of color vision
- Optic disc edema
- Optic atrophy
Optic nerve diseases:
Optic nerve diseases:

- **Typical optic neuritis:**
  - Inflammatory demyelinating condition associated with MS
  - Most common type in young adults
  - Good recovery
  - IV steroids may speed up the recovery process but does not influence the final outcome.
Optic nerve diseases:

- **Ischemic optic neuropathy (ION):**
  - *Non-arteritic ION:*
  - Patients often have DM, HTN and other vascular risk factor.
  - Most common cause in older patients
  - Altitudinal visual field loss
Ischemic optic neuropathy (ION):

- **Arteritic ION:**
  
  - >55yrs old
  
  - Associated with giant cell arteritis (GCA)
  
  - Check for jaw claudication, proximal myalgia and arthralgia, scalp tenderness, headache
  
  - Elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)
Optic nerve diseases:

- **Arteritic ION:**
  - Temporal artery biopsy is the gold standard for diagnosis
  - Treatment: Systemic steroids is given immediately if GCA is suspected
  - Binocular involvement occurs in third of cases, often within the first day.
Optic nerve diseases:

**Congenital disc elevation: <1%**

- Optic disc margins blurred and the cup is absent but no edema or hrg can be observed.
- May be associated with hyperopia or drusen
Other causes of optic neuropathy:

- Infection e.g. viruses, TB, cryptococcus and syphilis
- Systemic connective tissue disease e.g. SLE
- Genetics: Leber’s optic neuropathy (through a mitochondrial DNA mutation)
- Toxic and nutritional deficiencies
- Trauma
Papilledema

- Bilateral swelling of the optic discs secondary to increased intracranial pressure
Papilledema

- Hyperemia of the disc
- Tortuosity of the veins and capillaries
- Blurring and elevation of disc margins
- Peripapillary flame shaped haemorrhages
Papilledema

Look for spontaneous venous pulsations
Papilledema

Causes:
- Intracranial mass
- Sever systemic hypertension
- Idiopathic intracranial hypertension (pseudotumor cerebri)
a patient presented with this visual field defect.

Which one of the following diagnosis is the most likely?

a. Optic neuritis
b. tilted discs
c. pituitary tumor
d. 6th nerve palsy
Thank you